Metachronous Multiplicity of Spinal Cord Arteriovenous Fistula and Spinal Dural AVF in a Patient with Hereditary Haemorrhagic Telangiectasia

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Summary

HHT (Hereditary Haemorrhagic Telangiectasia or Rendu Osler Weber disease) is a known autosomal dominant dysplasia. The first clinical presentation of HHT in a child may be a cerebral or spinal AVM. We present the case of a young boy with HHT who had a previous spinal cord AVF treated by surgical obliteration and then presented with a spinal dural AVF nine months later.

This patient had surgical obliteration of a spinal cord perimedullary AVF and subsequently developed a new spinal dural AVF at a different level. The diagnosis was made by spinal MR imaging and spinal angiography

Introduction

HHT (Hereditary Haemorrhagic Telangiectasia or Rendu Osler Weber disease) is a known autosomal dominant dysplasia with epidemiological prevalence around 1/ 8,000 births [4]. The prevalence varies and may range from 1/3,500 to 1/5,000 in specific regions [19]. The phenotype is often restricted to some specific organ vessels including the nose, skin, lung, GI tract, liver and CNS (cerebral or spinal AVMs). To establish a diagnosis of HHT, at least two criteria must be met among which are: recurrent epistaxis, typical mucocutaneous telangiectasia, positive family history of HHT with a first-degree relative affected, and a typical AVM [9]. They are uncommon in the pediatric population and reported CNS manifestations are even less frequent [11,20]. The first clinical presentation of HHT in a child may be a cerebral or spinal AVM [10] and there is a higher association between HHT and perimedullary AVFs [11,12].

Spinal dural arteriovenous fistulas (SDAVFs) are abnormal arteriovenous communications within or adjacent to the dura. We present the case of a young boy with HHT who had a previous spinal cord AVF treated by surgical obliteration and then presented with a spinal dural AVF nine months later.

Case Report

A 14-year-old boy with a known family history of HHT presented with bilateral lower limb paralysis and bladder dysfunction in 1996. The only history noted was occasional epistaxis. His presenting symptom was lower back pain worsening over a five day period and requiring admission to hospital. Neurological examination revealed flaccid bilateral lower limb paralysis, absent anal tone and decreased lower limb reflexes. A lumber puncture demonstrated SAH at that time. A perimedullary AVF at T11 was diagnosed by MRI and DSA and was treated surgically the next day (figure 1A, 1B). An

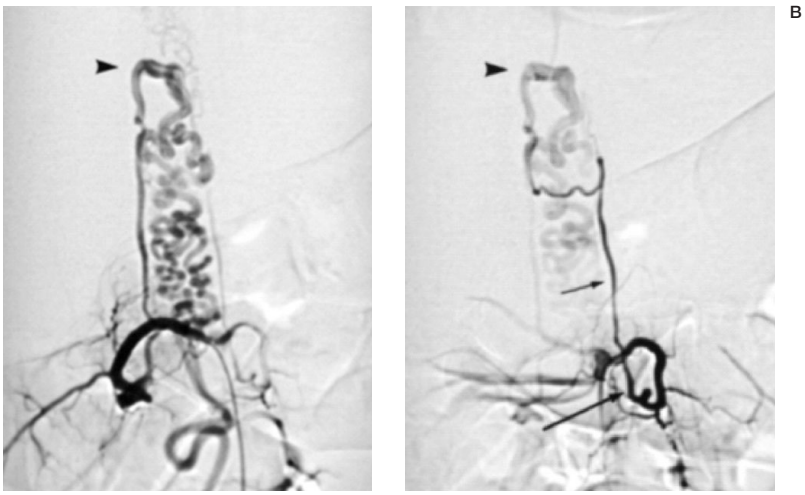

Figure 1 A,B) Spinal angiogram performed with contrast injection at L1 level showed posterior spinal arteries (arrow) converging towards the same posteriorly located perimedullary AVF (arrowhead).

intramedullary clot was identified and removed at surgery and the perimedullary AVF obliterated. He made an excellent recovery over approximately six months. He returned to full function with the exception of a mild right lower extremity weakness. Subsequently he gradually developed new bowel and bladder symptoms, which led to re-investigation. On examination, he had grade 4+ out of five strength of his right plantar flexion with increased tone, reflexes and an upgoing toe on the right, but was otherwise neurologically intact.

By the end of 1996, follow-up imaging was performed including MRI and angiography, which demonstrated the original lesion to be cured but a new fistula was discovered at the T12 level (figure 2A, 2B). The MRI of the T-spine at this stage showed a long tortuous signal void lesion at the level of T8-T12 with hyperintense T2WI signal changes compatible with cord edema from venous hypertension. This represented a change from the previous examination. Spinal angiography demonstrated a new spinal dural AVF at T12 on the left side, a level previously shown to be normal. Cure was achieved by a single injection of liquid adhesive resulting in obliteration of the fistulous communication and draining vein. His post-embolization course was uneventful and he remained neurologically stable on routine follow-up examinations for more than three years.

Discussion

Upon careful questioning, our case revealed a history of clinically insignificant epistaxis and

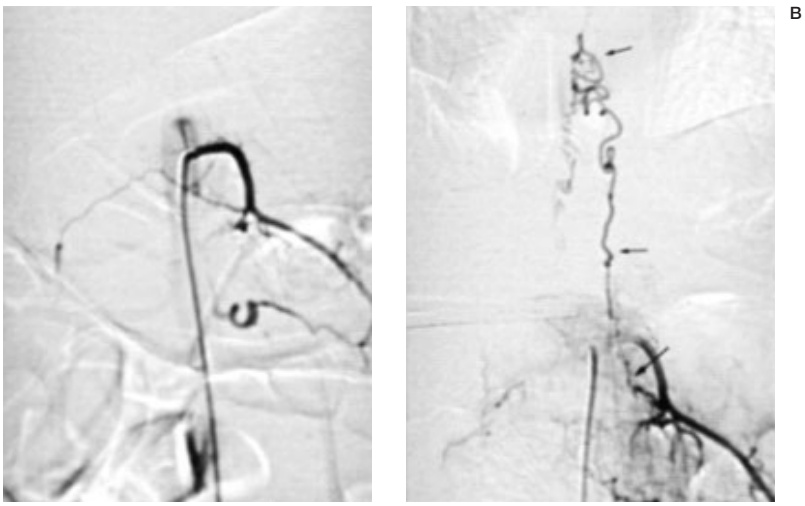

Figure 2 A) Post-OP spinal angiogram performed in March 1996 at left T12 level showed no evidence of fistula. B) There is a dural arteriovenous fistula (larger arrow) fed by arterial branches arising from the left T12 segmental vessels. Extensive venous reflux extending rostrally to the T8-9 disc level (smaller arrows) and then caudally back to the T12 level exiting on the side. None of the left posterior spinal artery at T10 level or the anterior spinal artery at L1 were participating the fistula.

during later childhood presented with a sudden onset of neurological deficit caused by a spontaneous SAH. Without any evidence of cerebral involvement, this finding should alert us to a possible spinal cause. Nevertheless, the diagnosis is made before the age of 15 in only 14% of spinal AVMs, with an additional 16% remaining undiagnosed till adulthood [15]. In children, spinal AVMs can cause severe back pain after a bleed within the cord similar to our case. Slowly progressive neurological manifestations are less common. Associated clinical findings can be scoliosis, abnormal gait, congestive heart failure, cutaneous vascular malformations and musculoskeletal asymmetry. Spinal AVMs generally become symptomatic during the second or third decade of life [2].

In children, a single clinical CNS manifestation may result in a failure to make an early diagnosis of HHT. Some research laboratories can provide a DNA mutation analysis for a definitive diagnosis. HHT can present with multiple pial AVFs and they may be the only manifestation of the disease [8]. Screening for pulmonary AVFs or brain AVFs or AVMs is recommended for family members. Brain abscess occurs in at least 5% of patients with HHT and PAVM [19].

There has been one documented case of spontaneous thrombosis of a spinal arteriovenous malformation following haemorrhage [5], but a metachronous spinal arteriovenous lesion in a patient with HHT has to our knowledge not been reported in the literature.

For the identification of spinal cord vascular lesions magnetic resonance imaging with contrast enhancement (ATECO) is more sensitive than MRA imaging in depicting residual or recurrent flow in peri- or intramedullary vessels and is recommended when clinical worsening occurs following initial improvement [6].

Perimedullary arteriovenous fistulae are usually found ventrally or dorsally along the surface of the spinal cord and are directly supplied by the anterior spinal and posterior spinal arteries. There is no intervening capillary network and the fistula does not penetrate the spinal cord. Venous drainage is into the coronal venous plexus.

Multiplicity of spinal arteriovenous fistulae, not HHT related, has been documented in one case [17]. Delayed post-surgical development of dural arteriovenous fistula can occur and was once reported in the literature [7]. The prior surgery may have created a wound with dural injury, which subsequently may have triggered angiogenetic factors and thus created a new AVF at dural level. The surgical exploration was higher up in our case at the upper thoracic level and is unlikely to be the direct cause for the newly developed dural fistula, which corresponds to a single, slow-flow direct AVF fed by non-dilated arterial branches of the left T12 segmental artery with a dilatation of the venous side of the shunt.

Spinal dural arteriovenous fistulas (SDAVF) are abnormal arteriovenous communications within the dura and perceived to be an acquired disorder. They are supplied by the branches of the vertebral, intercostal, lumbar, middle sacral or subclavian arteries or rarely by the branches of the internal iliac artery and can be purely fistulous, nidus type, a combination or part of a metameric lesion [3]. All purely fistulous malformations are perimedullary in location. In rare situations HHT type 1 patient giant perimedullary AVF can occur [12,18].

Myelopathy results from venous congestion, which can be eliminated by obliteration of the fistula. Spinal dural AVF is the most common variety of spinal vascular malformation. Atkinson et Al suggested the following clinical prognostic factors for patients who are expected to have a good functional prognosis after treatment at age below 70 years, Age below 70 years, less than 2.5 years after the onset of gait problem, slight or moderate incontinence and presence of the deep tendon reflexes in the lower extremities [1]. Van Dijk et Al 2003 found in the Toronto experience no particular criteria predicting favorable outcome [21].

Conclusions

We present the very rare metachronous occurrence of a perimedullary and a dural AVF in a young HHT patient. This patient had surgical obliteration of a spinal cord perimedullary AVF and subsequently developed a new spinal dural AVF at a different level. The diagnosis was made by spinal MR imaging and spinal angiography. The choice of treatment for the perimedullary AVF was guided by the perceived need for simultaneous evacuation of the spinal cord haematoma while endovascular intraarterial treatment was a good therapeutic option because of the favorable angioarchiecture of the spinal D-AVF.

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